

PATENT CLAIMS

5 1. An *in vitro* method of functionally determining at physiological conditions deficiencies in the lectin pathway of the complement system, employing a sample of mammalian blood, serum, plasma, or another body fluid obtained from a mammal, the method comprising the steps of

- 10 (a) adding an C1 complex inhibitor selected from the group consisting of proteins, peptides or immunoglobulins against C1q, C1r or C1s;
- (b) diluting the sample to inhibit the activation of the alternative pathway;
- 15 (c) adding a MBL or ficolin binding carbohydrate activating the lectin pathway in the sample;
- (d) adding an first antibody against the autologous C5b-9 complex and
- (e) determining the activation of the lectin pathway at the
- 20 physiological condition by measuring the autologous C5b-9 complex.

25 2. The method according to claim 1, wherein the inhibitor in step (a) is selected from the group consisting of C1 inhibitor, CRT, C1qr, E.coli C1g binding protein, gC1qR, ghB3, decorin, chondroitin sulphate proteoglycan, surfactant protein A and HNP-1.

30 3. The method according to claim 1, wherein the inhibitor in step (a) is selected from the group consisting of TDGDKAFVDFLSDEIKKEE, KDIRCKDD, AEAKAKA, VQVHNAKTKPR, WY, CEGPFGPRHDLTFCW and LEQGENVFLQATLL.

BEST AVAILABLE COPY

AMENDED SHEET

ART 34 AMDT

4. The method according to claim 1, wherein the inhibitor in step (a) is selected from the group consisting of polyclonal and monoclonal antibodies, such as IVIg and anti-Clq mAB 2204.

5. The method according to any of preceding claims, wherein the carbohydrate in step (c) is selected from the group consisting of mannose, fructose, mannan such as glucomannan and galactomannan, synthetic carbohydrate and microbial polysaccharide.

6. The method according to any of preceding claims, wherein the first antibody in step (d) is a polyclonal or a monoclonal antibody.

7. The method according to claim 6, wherein the step in (d) comprises adding a second antibody against the first antibody.

8. The method according to any of preceding claims, wherein the first or the second antibody is a labeled antibody.

9. A kit for functionally determining in a body fluid from a mammal deficiencies in the lectin pathway of the complement system, which kit comprises (a) an inert carrier and a MBL or ficolin binding carbohydrate (b) a diluent comprising a C1 complex inhibitor selected from the group consisting of molecules, peptides, proteases or immunoglobulins against Clq, C1r or C1s and (c) an first antibody against the autologous C5b-9 complex.

ART 34 AMDT

10. The kit according to claim 9, wherein the carbohydrate in (a) is selected from the group consisting of mannose, fructose, mannan such as glucomannan and galactomannan, synthetic carbohydrate and microbial polysaccharide.

11. The kit according to claim 9-10, wherein the inhibitor in (b) is selected from the group consisting of C1 inhibitor, CRT, C1qR, E.coli C1g binding protein, gC1qR, ghB3, decorin, chondroitin sulphate proteoglycan, surfactant protein A and HNP-1.

12. The kit according to claim 9-11, wherein the inhibitor in (b) is selected from the group consisting of the peptides, TDGDKAFVDFLSDEIKEE, KDIRCKDD, AEAKAKA, VQVHNAKTKPR, WY, CEGPFGPRHDLTFCW and LEQGENVFLOATLL.

13. The kit according to claim 9-12, wherein the inhibitor in (b) is selected from the group consisting of polyclonal and monoclonal antibodies, such as IVIg and anti-C1q mAB 2204.

14. The kit according to any of claims 9-13, wherein the first antibody in (c) is a polyclonal or monoclonal antibody.

15. The kit according to any of claims 9-14, wherein the carbohydrate in (a) is coated on the inert carrier.

16. The kit according to any of claims 9-15, wherein the first antibody in (c) is a labeled antibody.

ART 34 AMDT

17. The kit according to any of claims 9-16, wherein the kit further comprises a labeled second antibody (d) against the antibody in (c).

5

18. The kit according to claim 17, wherein the label is a fluoroscent or an enzyme label.

19. The kit according to claim 9-18, wherein the kit further comprises an enzyme substrate (e).

10

20. The kit according to any of claims 9-19, wherein the kit further comprises a washing solution (f).

15

21. The kit according to any of claims 9-20, wherein the kit further comprises a normal body liquid from a mammal (g).

22. The kit according to claim 21, wherein the normal body liquid (g) is a human serum.

20

23. The kit according to any of claims 10-23, wherein the kit further comprises an inactivated normal body liquid from a mammal (h).

25

24. The kit according to claim 24, wherein the inactivated normal body liquid (h) is heat inactivated human serum.

30